



## AMENDMENTS

### IN THE CLAIMS

Please cancel claims 11, 20-28, 30 and 34. Please amend claims 1, 2 and 33 as shown below.

1. (Currently Amended) A standardized prion preparation, comprising:  
prions obtained from a plurality of transgenic mouse brains; and  
a carrier;  
wherein the preparation comprises prions (a) which infect and cause disease in an animal chosen from a human, a cow, and a sheep, (b) which are prions of a known strain, (c) the prions are present in a known number of infectious units, and further wherein the carrier is different from brain tissue of the animal chosen from a human, a cow and a sheep.
2. (Currently Amended) The preparation of claim 1, wherein the prions are present in a ~~known number of infectious units and~~ a known concentration.
3. (Previously Presented) The preparation of claim 1, wherein the preparation is comprised essentially of transgenic mouse brain homogenate.
4. (Previously Presented) The preparation of claim 1, wherein the prions are of a single strain.
5. (Original) The preparation of claim 4, wherein the strain has a polymorphism selected from the group consisting of: human M129, human V129, human E219, human K219, sheep R171, sheep E171, sheep A136, sheep V136, a bovine 5 octarepeat polymorphism, and bovine 6 octarepeat polymorphism.
6. (Original) The preparation of claim 4, wherein the strain is human and has a pathogenic mutation selected from the group consisting of: a 2 octarepeat insert, a 4 octarepeat insert, a 5 octarepeat insert, a 6 octarepeat insert, a 7 octarepeat insert, an 8 octarepeat insert, a 9 octarepeat

insert, P102L, P105L, A117V, D178N, V180I, F198S, E200K, V210I, D217R, M232A, and a stop codon at 145.

7. (Original) The preparation of claim 1, wherein the prions are of a plurality of different known strains and wherein the prions are obtained from 10 or more animals, and further wherein the prions are produced in a transgenic mouse having a genome comprising exogenous genetic material encoding at least a portion of a PrP protein.

8. (Previously Presented) The preparation of claim 1, wherein the prions are obtained from a mouse having an ablated endogenous PrP gene and a genome manipulated to express a high copy number of an exogenous PrP gene from a genetically diverse animal chosen from a human, a cow and a sheep, and wherein the mouse spontaneously forms prions that normally infect the genetically diverse animal.

9. (Previously Presented) The preparation of claim 1, wherein the prions are uniformly dispersed in the preparation and are produced in a transgenic mouse selected from the group consisting of: Tg(HuPrP), Tg(HuPrP)/Prnp<sup>+/-</sup>, Tg(HuPrP)/Prnp<sup>0/0</sup>, Tg(HuPrP<sup>CJD</sup>), Tg(HuPrP<sup>CJD</sup>)/Prnp<sup>+/-</sup>, Tg(HuPrP<sup>CJD</sup>)/Prnp<sup>0/0</sup>, Tg(ShePrP), Tg(ShePrP)/Prnp<sup>+/-</sup>, Tg(ShePrP)/Prnp<sup>0/0</sup>, Tg(BovPrP), Tg(BovPrP)/Prnp<sup>+/-</sup>, and Tg(BovPrP)/Prnp<sup>0/0</sup>.

10. (Original) The preparation of claim 1, produced in a transgenic mouse selected from the group consisting of: Tg(MHu2M), Tg(MHu2M)/Prnp<sup>+/-</sup>, and Tg(MHu2M)/Prnp<sup>0/0</sup>.

11. – 17. (Canceled)

18. (Previously Presented) The preparation of claim 1, comprising between 0.1 and about 100 infectious units of prions.

19. (Previously Presented) The preparation of claim 1, comprising between about 1 and about 10 infectious units of prions.

20. – 28. (Canceled)

29. (Previously Presented) The preparation of claim 1, wherein the transgenic mouse brains are obtained from a transgenic mouse having both endogenous PrP genes ablated.

30. (Canceled)

31. (Previously Presented) The preparation as claimed in claim 1, wherein the prions infect and cause disease in a human.

32. (Previously Presented) The preparation of claim 1, wherein the transgenic mouse brains are produced in transgenic mice which are Tg(BovPrP)/Prnp<sup>0/0</sup>.

33. (Currently Amended) A standardized prion preparation, comprising:

prions obtained from a plurality of mice which are Tg (BovPrP)Prnp<sup>0/0</sup>; and  
a carrier;

wherein the prions are present in a known number of infectious units infect and cause disease in a cow and are prions of a known strain.

34. (Canceled)